Localising and lateralising value of ictal piloerection

T Loddenkemper, C Kellinghaus, J Gandjour, D R Nair, I M Najm, W Bingaman, H O Lüders

Background: Piloection is a rare clinical symptom described during seizures. Previous reports suggested that the temporal lobe is the ictal onset zone in many of these cases. One case series concluded that there is a predominant left hemispheric representation of ictal cold. The aim of this study is to evaluate the localising and lateralising value of pilomotor seizures.

Methods: Medical records of patients who underwent video electroencephalogram (EEG) monitoring at the Cleveland Clinic between 1994 and 2001 were reviewed for the presence of ictal piloerection. The clinical history, physical and neurological examination, video EEG data, neuroimaging data, cortical stimulation results, and postoperative follow ups were reviewed and used to define the epileptogenic zone. Additionally, all previously reported cases of ictal piloerection were reviewed.

Results: Fourteen patients with ictal piloerection were identified (0.4%). Twelve out of 14 patients had temporal lobe epilepsy. In seven patients (50%), the ictal onset was located in the left hemisphere. Four out of five patients with unilateral ictal piloerection had ipsilateral temporal lobe epilepsy as compared with the ipsilateral side of pilomotor response. Three patients became seizure-free after left temporal lobectomy for at least 12 months of follow up. An ipsilateral left leg pilomotor response with simultaneously recorded after-discharges was elicited in one patient during direct cortical stimulation of the left parahippocampal gyrus.

Conclusions: Ictal piloerection is a rare ictal manifestation that occurs predominantly in patients with temporal lobe epilepsy. Unilateral piloerection is most frequently associated with ipsilateral focal epilepsy. No hemispheric predominance was found in patients with bilateral ictal piloerection.

RESULTS

Fourteen right-handed patients (ten males, four females) with ictal piloerection were found (table 1). None of these patients had ictal piloerection as the sole seizure manifestation. Ictal piloerection was documented in nine cases by observation and video recordings and in five cases by the patients’ history. In 12 cases (85%), the suspected epileptogenic zone was located in the temporal lobe as demonstrated either by EEG (three patients), neuroimaging findings (six patients), or by seizure freedom after temporal lobectomy (three patients). In the other two cases the location of the epileptogenic zone could not be defined with certainty because of seizure recurrence in spite of temporal lobectomy.

In seven patients (50%), the ictal EEG onset was located in the right hemisphere and one patient had pilomotor seizures with ictal EEG seizure patterns arising independently from the left and right hemisphere. Nine patients (64%) experienced bilateral ictal piloerection and five patients had unilateral (or initially unilateral) ictal piloerection. Five out of the nine patients (56%) with bilateral piloerection had right temporal lobe epilepsy. Four out of the five patients (80%) with unilateral (or initially unilateral) ictal piloerection had the ictal onset in the hemisphere ipsilateral to the side of piloerection. Five patients underwent left and two patients right temporal lobectomy. Three patients became seizure free for at least 12 months of follow up, in two patients follow up is pending, and two patients continue to have seizures 2 and 6 months after surgery but only one of...
### Table 1: Fourteen patients with ictal piloerection

<table>
<thead>
<tr>
<th>Age, sex, handedness</th>
<th>Related condition</th>
<th>Seizure semiology (frequency)</th>
<th>Awareness of pilo</th>
<th>Distribution of pilo</th>
<th>EEG</th>
<th>Imaging findings</th>
<th>Surgery, outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>36, M, R</td>
<td>None</td>
<td>Olfact/gust aura — automotor (LOC) — pellag/parathesias, L pilo (1/d)</td>
<td>No</td>
<td>L body, ipsilateral</td>
<td>Interictal: LT 90%, RT 10%</td>
<td>MRI: normal</td>
<td>–</td>
</tr>
<tr>
<td>41, M, R</td>
<td>Head trauma</td>
<td>HV/pilo/diaphoresis (evolves into GTC) (3/d)</td>
<td>Yes</td>
<td>Bilateral</td>
<td>Ictal: LT</td>
<td>MRI: normal</td>
<td>–</td>
</tr>
<tr>
<td>51, M, R</td>
<td>None</td>
<td>Cold, pilo — nausea, SOB, loneliness, distress, tachycardia (1–2/d)</td>
<td>Yes</td>
<td>Bilateral</td>
<td>Ictal: LT</td>
<td>MRI: R HA</td>
<td>–</td>
</tr>
<tr>
<td>15, F, R</td>
<td>TS, mild DD</td>
<td>Cold, pilo, nausea — LOC, automatisms (→ GTC) (1–2/d)</td>
<td>Yes</td>
<td>Bilateral</td>
<td>Ictal: RT</td>
<td>MRI: R HA and multiple tubers</td>
<td>RT, seizures returned 2 mo after surgery (1/mo)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>S/p RT lobectomy</td>
<td></td>
<td></td>
<td>Ictal: RT</td>
<td>PET: RT hypometabolism</td>
<td>MRI: S/p RT lobectomy, multiple bil hamartomas</td>
</tr>
<tr>
<td>27, M, R</td>
<td>None</td>
<td>L arm pilo, cold, diaphoresis, SOB — automatisms (LOC) — GTC (2/week)</td>
<td>Yes</td>
<td>Initial L body, ipsilateral</td>
<td>Interictal: LT 75%, RTP 25%</td>
<td>MRI: cavernous angioma L superior T gyrus</td>
<td>L superior T, with intraoperative language mapping, seizure free</td>
</tr>
<tr>
<td>21, M, R</td>
<td>None</td>
<td>Hyperventilation, gust aura, hyperperspiration, pilo — LOC, automatisms, hyperlacrimation (→ GTC) (3/week)</td>
<td>No</td>
<td>Bilateral</td>
<td>Ictal: LT</td>
<td>PET: LT hypometabolism</td>
<td>MRI: L T MCD</td>
</tr>
<tr>
<td>57, M, R</td>
<td>None</td>
<td>Paresthesias in the nose, light-headed, bilateral arm and leg pilo (2/week)</td>
<td>Yes</td>
<td>Bilateral</td>
<td>Interictal: none</td>
<td>PET: L – RT hypometabolism</td>
<td>MRI: R HA</td>
</tr>
<tr>
<td>24, F, R</td>
<td>Head trauma age 8 mo</td>
<td>Two seizure types were recorded: 1. warning (strange feeling), perioral paresthesias, staring, LOC, automatisms (3/month) 2. staring, LOC, pilo R postictally (1/week)</td>
<td>No</td>
<td>R body, ipsilateral</td>
<td>Interictal: LT 95%, LT 5%</td>
<td>MRI: L HA</td>
<td>–</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ictal: LT</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>53, M, R</td>
<td>Head trauma age 36</td>
<td>Ab dura, pilo both arms (GTC) (5–6/d)</td>
<td>Yes</td>
<td>Initial L body, ipsilateral</td>
<td>Interictal: RT</td>
<td>MRI: R HA</td>
<td>–</td>
</tr>
<tr>
<td>21, M, R</td>
<td>Meningitis age 1 month, febrile convulsions age 1 mo</td>
<td>Pilo L leg, then bil, cephalic sensation, staring, LOC, hand automatisms, R arm stiffening, occasional postictal aphasia (1/d)</td>
<td>Yes</td>
<td>Initial L body, ipsilateral</td>
<td>Interictal: LT</td>
<td>MRI: L HA and L FP encephalomalacia</td>
<td>LT, seizure free</td>
</tr>
<tr>
<td>45, M, R</td>
<td>Head trauma in childhood</td>
<td>Ab d and olf aura, light-headed, No difficulties understanding, bil pilo (15/6)</td>
<td>No</td>
<td>Bilateral</td>
<td>Interictal: LT</td>
<td>MRI: LH enlargement, enhancement of LH head after gald Path: CNS-vasculitis</td>
<td>MRI: normal</td>
</tr>
<tr>
<td>49, M, R</td>
<td>None</td>
<td>Cephalic sensation, anxiety, tachycardia, pilo (1–2/d)</td>
<td>No</td>
<td>Bilateral</td>
<td>Interictal: RT</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>53, F, R</td>
<td>Low grade glioma</td>
<td>Urinary urge, fear, tachycardia, HV, abd aura, bil pilo, chills, L face tonic, GTC (1/10)</td>
<td>Yes</td>
<td>Bilateral</td>
<td>Ictal: RT</td>
<td>MRI: R FT low-grade glioma</td>
<td>LT, last to follow up</td>
</tr>
<tr>
<td>54, F, R</td>
<td>AVM rupture</td>
<td>HV, chills, fear, tachycardia, shaking, L arm pilo, then bil, urinary urge, automatisms, no LOC (3–4/mo)</td>
<td>Yes</td>
<td>Initial L body, contralateral</td>
<td>Interictal: RT 90%, RTP 10%</td>
<td>MRI: R superior T encephalomalacia after AVM rupture</td>
<td>RT, follow up pending</td>
</tr>
</tbody>
</table>

→, evolves into; A, atrophy; abd, abdominal; AVM, arteriovenous malformation; bil, bilateral; CNS, central nervous system; d, day; DD, developmental delay; EEG, electroencephalogram; F, female; GAD, gadolinium; GTC, generalised tonic-clonic seizure; gust, gustatory; H, hippocampal; HV, hyperventilation; IRS, intermittent rhythmic slow; L, left; LOC, loss of consciousness; M, male; MCD, malformation of cortical development; mo, months; MRI, magnetic resonance imaging; olfact, olfactory; R, right; SOB, shortness of breath; T, temporal; O, occipital; P, parietal; PET, positron emission tomography; pilo, piloerection; S/p, status post; SPECT, single photon emission computed tomography; TS, tuberous sclerosis; yr, year.
in the mesial frontal or orbital frontal plates. The EEG seizure onset started from the two most mesial electrodes of the basal temporal plates.

During direct cortical stimulation of the left parahippocampal gyrus, a pilomotor response in the ipsilateral left leg was observed simultaneously with an after-discharge confined to the stimulated electrode. A left anterior temporal lobectomy was performed and histology revealed malformation of cortical development. Rare seizures returned 6 months after surgery, but seizure semiology changed and did not include piloerectio postoperatively (table 1).

**DISCUSSION**

Ictal piloerectio occurs predominantly in patients with temporal lobe epilepsy. This is consistent with previously reported cases as evidenced by EEG and imaging.4,5,6,7 Only a few exceptions have been described with seizures arising from the frontal,8–11 parieto-occipital,12 fronto-parietal,13 and fronto-temporal14 brain regions.

Although a previous case series found left hemispheric epilepsy in 18/26 patients (69%) with ictal cold shiver (five patients), ictal piloerectio (nine patients), or both (12 patients),9 the lateralisng value of ictal piloerectio seems to be less clear. In our series, six out of 14 (43%) patients had left hemispheric epilepsy. In 23/38 (61%) of reported patients with ictal piloerectio in whom the lateralisng information was provided, the epileptogenic zone was most likely in the left hemisphere.4–7,11,13,16,22–27 Therefore, ictal piloerectio in general seems to have no lateralisng value. The left hemisphere predominance in the series of Stefan et al.12 may be related to the inclusion of patients with the feeling of ictal cold.

In contrast, unilateral (or initially unilateral) ictal piloerectio is usually associated with ipsilateral seizure onset. Four of our five patients with unilateral onset of ictal piloerectio had ipsilateral focal epilepsy. Review of the literature revealed 18 cases with clearly unilateral (or initially unilateral) ictal piloerectio (table 2).4–7,9–11,13,14,16,22–27 In 14 of these patients, the epileptogenic zone could be lateralisng. Twelve of these 14 patients (86%) had an epileptogenic zone that was in the hemisphere ipsilateral to the side of ictal piloerectio as documented by successful epilepsy surgery,4–7,9–11,13,14,16,22–27 and by imaging and EEG findings.4–7,9–11,13,14,16,22–27 In four patients, the epileptogenic zone could not be lateralisng.4–7,13,16,22–27 Only two previously reported cases and one of our cases showed ictal piloerectio contralateral to the (probable) hemisphere of seizure onset.4–7,22 In summary, we found 19 patients with unilateral ictal piloerectio in whom the epileptogenic zone could be lateralisng. In 16 of these patients (84%), ictal piloerectio was ipsilateral to the epileptogenic zone.

In our patients, ictal piloerectio was never the only seizure manifestation. This corresponds with descriptions of ictal piloerectio in conjunction with an epigastric sensation,4–7,9–11,13,14,22–27 feelings of fear or other experiential sensations,4–7,9–11 olfactory hallucinations,4–7,10,13,27 unspecified warning sensations,4 or localised tingling.11,13,16,27 Additional autonomic features in the literature include descriptions of shivering or vasomotor symptoms in the literature.4–7,9–11,13,16,22–27 We cannot exclude the possibility that piloerectio could be secondary to psychic auras with anxiety or fear, or secondary to fear of an impending seizure. However, ictal piloerectio was the first seizure symptom in five out of 25 patients with detailed seizure descriptions.4–7,15,23,24,26,27 In some of our patients there were other clinical symptoms regularly following the piloerectio—for example,
Piloerection has been elicited by electrical or pharmacological stimulation without or only focal after-discharge. Electrical stimulation of the left parahippocampal gyrus produced an ipsilateral pilomotor response with ipsilateral piloerection. However, piloerection may be secondarily mediated, and not directly elicited by mesial temporal stimulation.

In one patient, subdural EEG recordings gave evidence for ictal onset in the cingulate gyrus. Additionally, electrical stimulation of the rostral cingular cortex in monkeys resulted in generalised piloerection. However, because of a lack of documentation of after-discharges, piloerection because of spread of the initial discharge cannot be excluded in this case.

Ipsilateral piloerection was also seen after electrical stimulation of the posterior hypothalamic region. This area is closely connected with the hippocampus through the fornix. The study was also not controlled for after-discharges.

Because of the fact that patients are not always aware of ictal piloerection and because of the fact that not all video recordings are sensitive enough to detect ictal piloerection we may have missed patients with piloerection, which may have lead to a sampling bias in our retrospective study. Prospective investigation of this ictal phenomenon with additional monitoring devices—for example, skin resistance testing—may improve the sensitivity to pick up ictal piloerection in future studies.

In summary, ictal piloerection is rare, associated with temporal seizure onset, without clear localising value if the piloerection is bilateral. However, if the piloerection starts unilaterally, the seizure onset is most likely lateralised to the ipsilateral hemisphere. The exact location of the generator of piloerection remains unclear. It is most likely situated within the central autonomic network possibly in the insula.
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